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Long-term Outcome of Optic Nerve Encasement and Optic Nerve Decompression in Patients with Fibrous Dysplasia: Risk Factors for Blindness and Safety of Observation

OBJECTIVE: Fibrous dysplasia (FD) of bone may occur solely as a skeletal condition or it may occur in association with extraskeletal manifestations, including growth hormone (GH) excess. Uncertainty exists as to the management of FD involving the optic nerves. In an effort to clarify management, the authors studied a large population of patients.

METHODS: One hundred four patients underwent an evaluation that included review of records, endocrine testing, cranial computed tomography, and neuro-ophthalmological examination.

RESULTS: Ninety-one of 104 patients had craniofacial FD; complete records were available for 87 patients (174 nerves). Seventeen percent of the optic nerves were less than 50% encased, 22% were 50 to 99% encased, and 61% were 100% encased. Twelve percent of the nerves that were 100% encased showed evidence of optic neuropathy, but 88% did not. The group with optic neuropathy was not older than the group without. Patients with GH excess were significantly more likely to have nerves that were 100% encased (relative risk, 4.1; 95% confidence interval, 1.5–11.1; $P = 0.0017$) and to have optic neuropathy (relative risk, 3.8; 95% confidence interval, 2.0–7.1; $P = 0.0019$). Six prophylactic optic nerve decompressions were performed; in five patients, vision was stable after surgery, and one patient was blind after surgery. Thirteen interventional optic nerve decompression procedures were performed; six of the 13 patients showed some improvement and seven of the 13 showed no improvement or worsened vision.

CONCLUSION: The vast majority of optic nerves encased with FD do not exhibit symptoms of optic neuropathy and seem to be stable over time. GH excess is associated with increased risk of nerve encasement and optic neuropathy. Patients with craniofacial FD should be screened for GH excess, and optic nerve decompression should be performed only when there is objective evidence of progressive optic neuropathy.

KEY WORDS: Bone, GNAS, Growth hormone, $G_s\alpha$, McCune-Albright syndrome

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Fibrous dysplasia (FD) of bone is a benign skeletal disease in which normal bone is replaced by a benign fibro-osseous tissue (5, 6, 15). It results from postzygotic activating mutations in the signaling protein $G_s\alpha$ (17). It may appear as a condition of the skeleton only or as part of the McCune-Albright syndrome (MAS), which is clinically defined by FD in combination with either café au lait skin pigmentation and/or at least one of a number of hyperfunctioning endocrinopathies, including precocious puberty, hyperthyroidism, growth hormone (GH) excess, and others (4). The craniofacial structures and cranial base are involved in many patients, and the optic nerves are usually involved and are often encased circumferentially as they pass through the cranial base (6, 8, 14). The optimal management of optic nerves circumferentially encased with fibrous dysplastic bone, but without symp-

toms of optic neuropathy, is controversial (3, 9–11, 13, 16). Prophylactic decompression is sometimes performed based on the assumption that the risk of future optic neuropathy outweighs the risks of the operation (3, 7, 10–13), which include postoperative blindness (3, 7, 13). The reason for this controversy is the lack of knowledge of the natural history and the risk for blindness in the absence of intervention. It has been pointed out that this controversy could be resolved by data on the natural history of this condition (9).

We previously demonstrated that significant narrowing of the optic canal with FD was not associated with optic neuropathy (8) and that GH excess may be related to a more severe craniofacial phenotype (1, 8, 19). Herein, we report on a larger group, with a longer follow-up period, who underwent a uniform and comprehensive evaluation. The goal was to better define the natural history of FD encasing the optic nerve and to identify the pathophysiological mechanisms contributing to the development of optic neuropathy.

PATIENTS AND METHODS

All patients seen at the National Institutes of Health since 1998 with a diagnosis of FD were evaluated. The diagnosis of FD was made based on the results of clinical, radiographical, and histological studies. Craniofacial FD was identified by a combination of nuclear medicine bone scans and computed tomographic (CT) analysis. All patients underwent testing of all relevant endocrine axes. A diagnosis of GH excess was made on the basis of a serum GH level of more than 1.0 ng/ml measured 60 minutes after a standard oral glucose tolerance test. Patients without neuro-ophthalmological examination results were excluded from the relevant sections of analysis. All patients were enrolled in an institutional review board-approved protocol and gave informed consent.

All patients were evaluated by a single neuro-ophthalmologist (EJF). Testing included best-corrected visual acuity, according to the Early Treatment Diabetic Retinopathy Study scale (20/20 denotes perfect vision); visual fields obtained using the Humphrey Visual Field Analyzer (Humphrey Instruments, San Leandro, CA) using the Swedish Interactive Thresholding Algorithm (SITA) 30–2 program or Goldmann perimetry testing; color vision, with the use of 14 Ishihara color plates; contrast sensitivity testing using the Pelli Robson charts; and results of examination of the fundus. Because there is no definitive test for optic neuropathy, abnormalities suggestive of optic neuropathy resulting from FD were defined as either an abnormal result on the visual field test (such as scotoma or field deficit) or an abnormal result on two of the three other tests performed (visual acuity less than 20/40, correct identification of fewer than 10 of 14 Ishihara color plates, or evidence of optic atrophy on examination of the fundus).

All patients underwent standardized CT imaging of the cranium, on either a 4- or 8-slice helical scanner, using 2.5- to 3.8-mm collimation. The slice reconstruction interval was 1.25 to 1.50 mm. Soft tissue and bone algorithm reconstructions were reviewed by a single neuroradiologist (JAB). The extent of encasement of optic nerves by FD of bone was evaluated in a semiquantitative manner as less than 50, 50 to 99, and 100%. Statistical analyses were performed using InStat software, version 3 (GraphPad Software, Inc., San Diego, CA).

RESULTS

One hundred four patients with FD and MAS were seen at the National Institutes of Health between 1998 and 2005. Of these, 91 (88%) had craniofacial involvement. Demographics for the group are shown in *Table 1*. Cranial CT scans were available for 91 patients. Neuro-ophthalmological evaluations were available for 87 (96%) of these 91 patients. In two patients, no examination was performed, and, in two patients, the record of the examination was missing. These four patients were excluded from the relevant analyses. Of the four patients excluded from the analysis because of no neuro-ophthalmological examination, four of the optic nerves were 100% encased and four were less than 50% encased with FD of bone.

TABLE 1. Patient demographics[a]

	Craniofacial fibrous dysplasia patients (n = 91)	Patients with 100% optic nerve encasement (n = 61)[b]	Patients with 50 to 99% optic nerve encasement (n = 28)[b]	Patients with <50% optic nerve encasement (n = 24)[b]
Age (yr)[c]				
Average	24.3	22.5[d]	25.7[d]	29.8[d]
Median	19	18	12.5	31
Range	3–84	3–84	6–84	6–69
Sex				
Male	39/91 (43%)	27/61 (44%)	11/28 (39%)	13/24 (54%)
Female	52/91 (57%)	34/61 (56%)	17/28 (61%)	11/24 (46%)
Diagnosis				
MFD	1/91 (1%)	0/61 (0%)	0/28 (0%)	1/24 (4%)
PFD	7/91 (8%)	0/61 (0%)	4/28 (14%)	5/24 (21%)
MAS	83/91 (91%)	61/61 (100%)	24/28 (86%)	18/24 (75%)

[a] MFD, monostotic fibrous dysplasia; PFD, polyostotic fibrous dysplasia; MAS, McCune-Albright syndrome, defined as fibrous dysplasia plus café au lait hyperpigmentation, endocrinopathies, or both.
[b] Some patients may fall into two categories.
[c] Age at date of most recent head computed tomographic scan.
[d] Age differences between groups not statistically significant ($P = 0.16$).

Optic Nerve Encasement

Of the 174 optic nerves, 29 (17%) were less than 50% encased, 38 (22%) were 50 to 99% encased, and 107 were 100% encased with FD of bone (*Fig. 1*). The average age of patients with optic nerves 100% encased was 22.5 years (median, 18 yr; range, 3–84 yr), compared with 25.7 years (median, 12.5 yr; range, 6–84 yr) and 29.8 years (median, 31 yr; range, 6–69 yr) for those with a nerve 50 to 99% encased or less than 50% encased, respectively (*Table 1*). There were no significant age differences between groups with less than 50, 50 to 99, and 100% encasement ($P = 0.16$, Krushkal-Wallis nonparametric analysis of variance), demonstrating a lack of age-related progression. Patients with GH excess were more likely to have optic nerves 100% encased by FD of bone than those without (relative risk, 4.1; 95% confidence interval, 1.5–11.1; $P = 0.0017$). All patients with at least one nerve 100% encased had MAS.

Optic Neuropathy

There was no evidence of optic neuropathy in any of the nerves that were either less than 50% or 50 to 99% encased by FD of bone (*Fig. 1*; *Table 2*). Of the nerves 100% encased by FD of bone, 94 (88%) of 107 exhibited no evidence of optic neuropathy. Optic neuropathy was seen in 13 (12%) of 107 nerves that were 100% encased. Of these, 12 (92%) were surgically decompressed and are discussed below. The remaining one (7%) had only mild neuropathy characterized by a pale fundus and a slow color vision response (10 out of 14 Ishihara color plates correct), which was not noticeable to the patient. This patient's mild optic neuropathy has been stable during 7 years of follow-up. The average age of patients at the time optic neuropathy occurred was 16.3 years (median, 12 yr; range, 5–35 yr), and the average age of those without optic neuropathy was 20.9 years (median, 18 yr; range, 3–84 yr). There was no age difference between the group with and without optic neuropathy ($P = 0.3057$, two-tailed Mann-Whitney *U* test).

Growth Hormone Excess and Aneurysmal Bone Cysts

Seven (54%) out of 13 nerves with optic neuropathy occurred in patients with GH excess (*Fig. 1*; *Table 2*). As such, GH excess represented a statistically significant risk factor for the development of optic neuropathy (relative risk, 3.8;

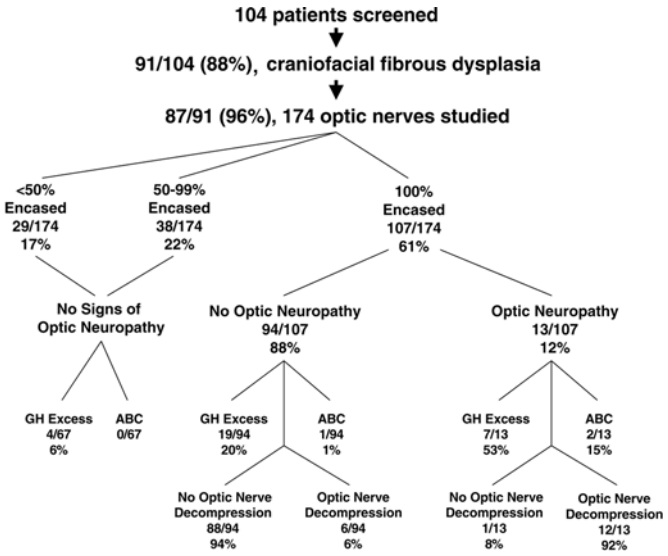

FIGURE 1. *Flowchart showing progress of patients studied. One hundred four patients were screened; 91 patients had craniofacial FD, and complete records were available for 87 patients (96%). The breakdown according to the extent of encasement and the presence or absence of GH excess, aneurysmal bone cyst (ABC), optic neuropathy, or a combination thereof are indicated. Optic neuropathy was defined as either an abnormal result on the visual field test or an abnormal result on two of the three other tests performed (visual acuity worse than 20/40, correct identification of fewer than 10 out of 14 Ishihara color plates, or evidence of optic atrophy on examination of the fundus).*

TABLE 2. Optic nerve encasement, optic neuropathy, growth hormone excess, and bone cysts[a]

	100% encasement	50–99% encasement	<50% encasement
No symptoms of optic neuropathy[b]	94/107 (88%)	38/38 (100%)	29/29 (100%)
Average age (yr)[c]	20.9	24.2	28.1
Median age (yr)	18	12.5	29
Age range (yr)	3–84	6–84	6–56
GH excess	19/94 (20%)	2/38 (5%)	2/29 (7%)
ABC	1/94 (1%)	0/38 (0%)	0/29 (0%)
Symptoms of optic neuropathy[d]	13/107 (12%)	0/38 (0%)	0/29 (0%)
Average age (yr)[c]	16.3 years	—	—
Median age (yr)	12 years	—	—
Age range (yr)	5–35 years	—	—
GH excess[e]	7/13 (54%)	—	—
ABC	3/13 (23%)	—	—

[a] GH, growth hormone; ABC, aneurysmal bone cyst. Preoperative extent of encasement and age at time that symptoms of optic neuropathy occurred were used for patients who had undergone optic nerve decompression.
[b] Optic neuropathy was defined by visual field defect or two of the following: decreased color vision, decreased visual acuity, abnormal appearance of fundus.
[c] Age differences between groups was not statistically significant ($P = 0.31$).
[d] One patient had optic nerve decompression of her right eye twice; both ages at times of optic nerve decompression were counted in these calculations.
[e] Patients with GH were more likely to have optic neuropathy (relative risk, 3.8; 95% confidence interval, 2.0–7.1; $P = 0.0019$).

95% confidence interval, 2.0–7.1; $P = 0.0019$). In two (15%) out of 13 nerves, the development of optic neuropathy was associated with an aneurysmal bone cyst (ABC) that compressed the optic nerves (*Fig. 1*), and one nerve (8%) was associated with both GH excess and an ABC. Therefore, in nine (69%) out of 13 nerves, optic neuropathy was associated with either GH excess or an ABC.

TABLE 3. Demographic data of patients who underwent optic nerve decompression[a]

	Patients without surgery (n = 79)	Surgery patients (n = 12)[b]	Interventional surgery (n = 9)[c]	Prophylactic surgery (n = 4)
Age (yr)[d]				
Average	24.6	14.8	14.8	15.8
Median	19	12	12	5.5
Range	3–84	5–37	5–30	5–37
Sex				
Male	35/79 (44%)	4/12 (25%)	3/9 (33%)	2/4 (50%)
Female	44/79 (56%)	8/12 (75%)	6/9 (66%)	2/4 (50%)
Diagnosis				
MFD	1/79 (1%)	0/12 (0%)	0/8 (0%)	0/4 (0%)
PFD	6/79 (8%)	1/12 (8%)	0/8 (0%)	1/4 (25%)
MAS	72/79 (91%)	11/12 (92%)	8/8 (100%)	3/4 (75%)

[a] MFD, monostotic fibrous dysplasia; PFD, polyostotic fibrous dysplasia; MAS, McCune-Albright syndrome, defined as fibrous dysplasia plus café au lait hyperpigmentation, endocrinopathies, or both.
[b] Age at date of most recent head computed tomographic scan.
[c] Age at date of surgery.
[d] One patient underwent interventional decompression on the same optic nerve at two different times; both ages at time of symptoms were included for calculation.

Optic Nerve Decompression

Optic nerve decompression was performed on 18 nerves. In the 94 patients without optic neuropathy, there were six prophylactic decompression surgeries (6%). Twelve (92%) of the 13 patients with optic neuropathy underwent 13 decompression surgeries (*Fig. 1*). All patients who underwent optic nerve decompression had nerves that were 100% encased. The demographics of the patients who underwent optic neuropathy are shown in *Table 3*. The average age of patients who underwent an interventional optic nerve decompression was 14.8 years (median, 12 yr; range, 5–30 yr). The average age at decompression for those who underwent prophylactic procedures was 15.8 years (median, 5.5 yr; range, 5–24 yr), and the average age of patients who did not undergo optic nerve decompression was 24.6 years (median, 19 yr; range, 3–84 yr).

Optic nerve decompressions were performed 13 times on 12 nerves (one for recurrent symptoms). Vision improved as a result of five (38%) of these procedures. One symptomatic nerve had improved vision after the initial optic nerve decompression, but then optic neuropathy developed 10 years later. The symptoms also resolved after the second optic nerve decompression. This patient had associated GH excess. Slight improvement was seen in one (8%), no improvement was reported in four (31%), and vision loss was reported in two (15%) of 13 procedures (*Table 4*).

Six prophylactic optic nerve decompressions were performed on six nerves that were 100% encased, but in which there were no symptoms of optic neuropathy (*Fig. 1*; *Table 4*). Five (83%) out of six of these nerves were intact after surgery with no change in vision. However, one (17%) of the six sustained intraoperative injury resulting in blindness.

DISCUSSION

The proper handling of optic nerves encased by FD without symptoms of neuropathy has long been controversial. Many authors recommend prophylactic decompression based on the assumption that the disease is progressive and that optic neuropathy is an inevitable complication. This recommendation assumes that the risk of prophylactic decompression—which includes blindness—is less than the risks associated with no treatment for asymptomatic nerves. In a previous study of 67 nerves, we demonstrated that narrowing of the optic canal alone is not necessarily associated with visual loss (8). In this study, we extend these findings to a larger cohort and demonstrate that, in most patients, vision loss is associated with either GH excess or an ABC.

In 93% of the 174 nerves, there was no evidence of optic neuropathy, and there was no optic neuropathy in nerves less than 100% encased. Of the 107 optic nerves that were 100% encased, 88% exhibited no evidence of optic neuropathy. There was no difference in age between groups with less than 50, 50 to 99, and 100% encasement, and patients with optic neuropathy were not older than those without optic neuropathy. These data support the conclusion that encasement does not progress with age and that increasing age does not necessarily bring with it the likelihood of optic neuropathy and blindness. Thus, prophylactic decompression should not be performed on patients without symptoms merely as an effort to prevent possible future optic neuropathy.

Two associations suggested the pathophysiological mechanism of optic neuropathy, namely GH excess and the presence of an ABC. Sixty-nine percent of the patients with optic neuropathy had either GH excess or an ABC of the cranial base. GH (and/or its trophic hormone insulin-like growth factor I) seems to promote growth and expansion of craniofacial FD. Patients with craniofacial FD and GH excess are more prone to macrocephaly, and there is a significant linear correlation between serum GH levels and head circumference in patients

TABLE 4. Optic neuropathy and optic nerve decompression: indications and outcomes[a]

Patient no.	Age (yr)	Eye	Indication at preoperative examination	Postoperative examination findings	Outcome summary	Postoperative encasement (%)	Follow-up (yr)	Endocrinopathies
Interventional optic nerve decompression (n = 9 patients; 12 nerves)								
1	24	R	VF	Moderate: CV; abnormal funduscopic results	Improved vision	100	18	GH excess, PP
2	5	L	VF	Mild: VF, CV; abnormal funduscopic results	Improved vision	100	12	HT, PW, neonatal Cushing's syndrome
3	14	R	VF, decreased VA	Mild: VF	Improved vision	100	25	GH excess, PP
3	24	R	VF, decreased VA, CV	Mild: VF	Improved vision	100	15	GH excess, PP
4	11	R	ABC, VF: vascular occlusion	Normal examination results	Improved vision	100	7	PP
5	12	R	ABC, decreased VA: light perception	Severe: VF, decreased VA, CV; abnormal funduscopic results: finger counting	Slight improvement	100	10	PP, HT
5	12	L	ABC, decreased VA: minimal light perception	Severe: VF, decreased VA, CV; abnormal funduscopic results: light perception, finger counting at 3–4 ft	No improvement	100	10	PP, HT
6	11	L	VF, decreased VA, CV	Moderate: VF, decreased VA, CV; abnormal funduscopic results	No improvement	100	2	PP
7	12	R	VF, decreased VA	Mild: VF, decreased VA	No improvement	100	1.5	GH excess, PP
8	30	R	VF, decreased VA, CV, abnormal funduscopic results	Severe: VF, decreased VA, CV; abnormal funduscopic results	No improvement	100	4	GH excess, PP, PW
3	14	L	Decreased VA	Severe: VF, decreased VA, CV; abnormal funduscopic results	No improvement	100	25	GH excess, PP
9	12	R	Decreased VA	Severe: VF, decreased VA, CV; abnormal funduscopic results	Severe vision loss	100	6	GH excess
9	12	L	Decreased VA	Severe: VF, decreased VA, CV; abnormal funduscopic results	Severe vision loss	100	6	GH excess
Prophylactic optic nerve decompression (n = 4 patients; 6 nerves)								
10	6	R	OC narrowing, right eye proptosis	Normal examination results	Normal visual results	50–99	3	None
11	37	R	OC narrowing: normal	Normal examination results	Normal visual results	<50	2	None
11	37	L	OC narrowing: normal	Normal examination results	Normal visual results	<50	2	None
12	5	R	OC narrowing: normal	Normal examination results	Normal visual results	100	9	GH excess, PP, HT
12	5	L	OC narrowing: normal	Normal examination results	Normal visual results	100	9	GH excess, PP, HT
2	5	R	OC narrowing: normal	Severe: VF, decreased VA, CV; abnormal funduscopic results	Vision loss	100	11	HT, PW, neonatal Cushing's syndrome

[a] R, right; VF, visual field defect; CV, decreased color vision; GH, growth hormone; PP, precocious puberty; L, left; HT, hyperthyroid; PW, phosphate wasting; VA, visual acuity; ABC, aneurysmal bone cyst; OC, optic canal. Data for indications for surgery were obtained by patient report or records. Postoperative examinations were performed by a neuro-ophthalmologist at our institution. Postoperative encasement of optical nerves with FD bone was determined by a neuroradiologist at our institution and was taken from the most recent head computed tomographic examination.

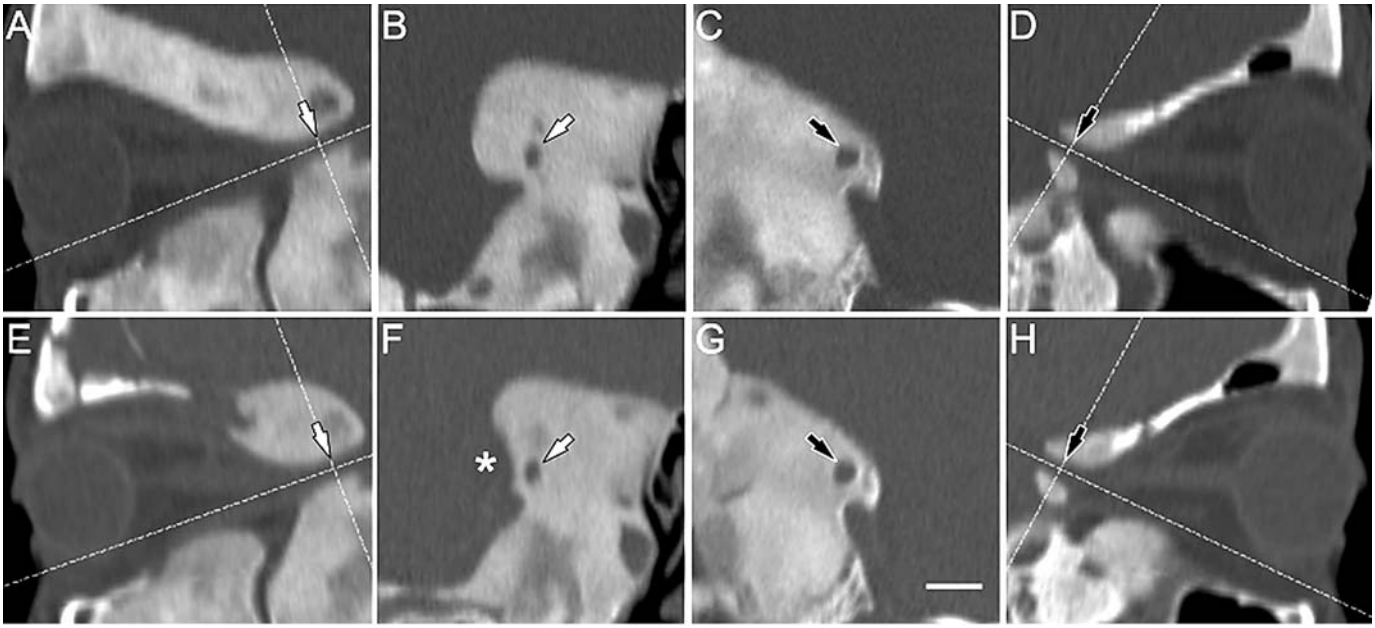

FIGURE 2. *Preoperative (*A–D*) and postoperative (*E–H*)CT scans obtained from a 12-year-old girl with MAS and GH excess (Patient 7,* Table 4*) who underwent optic nerve decompression for a mild visual field defect.* A–D, *preoperative images demonstrating full encasement of both nerves, with marked narrowing of the right optic canal (*A *and* B, white arrows*) as compared to the left (*C *and* D, black arrows*).* E *and* F, *postoperative images obtained 1.5 years after surgery demonstrating that some bone has been removed (*asterisk*), but the optic canal remains encased (*white arrows*).* G *and* H, *no change is noted on the left. There were essentially no differences in the pre- and postoperative neuro-ophthalmological examinations.* A, D, E, *and* H, *parasagittal oblique views along the plane of the optic canal.* B, C, F, *and* G, *coronal oblique views perpendicular to the long axis of the optic canal. Scale bar (*G*), 1 cm.*

with craniofacial FD and GH excess (2). Furthermore, there is evidence that optic nerve stretching resulting from bone expansion may be the mechanism of vision loss in some patients with GH excess and craniofacial FD (8). This is significant because GH excess is a potentially treatable disease (1, 18). Yet, early in the course of MAS, when intervention is perhaps most important to prevent long-term morbidity, GH excess is usually not clinically evident. Its presentation may be as subtle as normal stature in a young adult who experienced precocious puberty and should have short stature as a consequence of early growth plate closure. Therefore, referral for specific testing for GH excess is essential. This suggests that, in the absence of the comorbidities of GH excess, an ABC, or both, the baseline rate of optic neuropathy in craniofacial FD is quite low. With such a low rate of optic neuropathy, the risk of injury to the optic nerve during prophylactic optic nerve decompression becomes an even greater consideration.

Fifteen (83%) out of 18 patients who underwent optic nerve decompression still had 100% encasement of the optic nerve on postoperative cranial CT scans (*Fig. 2; Table 4*). It is not known whether this was the result of incomplete decompression or regrowth. Despite persistent (or recurrent) 100% encasement in these patients, only two out of 15 patients, both of whom had GH excess, experienced symptoms of optic neuropathy. This suggests that, even if FD of bone is removed from around the optic nerve, it is likely to recur, further questioning the prudence of decompression in the absence of symptoms.

It is possible that the low rate of optic neuropathy in the group is the result of referral bias because ours is a medically based treatment group. Against this argument is the fact that, as a group, this was a more severely affected group of patients with FD. The ratio of MAS to polyostotic FD to monostotic FD was 91:8:1, the relative inverse of a random group of patients with FD (6).

CONCLUSION

Complete encasement of the optic nerve in FD of the cranial base is common, but is not commonly associated with optic neuropathy. There does not seem to be an age-related progression to optic neuropathy in patients in whom the optic nerve is encased, suggesting that, in most cases, the condition is stable. The vast majority of cases of optic neuropathy are seen in patients with either GH excess, an ABC, or both. Risk factors for the development of ABCs are not known, but GH excess, which is seen in approximately 20% of the patients with MAS, is relatively easy to diagnose and is treatable. Patients with craniofacial FD should be screened for GH excess, and prophylactic decompression should be reserved for patients with objective signs of optic neuropathy.

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COMMENTS

The authors have presented an excellent retrospective review of their experience of more than 20 years in the management of patients with fibrous dysplasia of the cranial base. Clinical decision making in this group of patients is often difficult in terms of whether or not the optic nerves should be decompressed, when they should be decompressed, and how should they be decompressed. The authors' observations shed considerable light in these areas.

Importantly, they have demonstrated that, despite encasement of the optic nerves by dysplastic bone, the majority of patients do not develop symptoms of optic neuropathy and remain stable over time, except if there is an elevated growth hormone level or aneurysmal bone cyst. A cautionary note is raised in their results of six prophylactic optic nerve decompressions in asymptomatic patients. Although five out of six were intact postsurgically, one patient (17%) experienced blindness from the prophylactic procedure.

The authors fail to discuss their surgical technique for optic nerve decompression, but presumably this was via a transcraniotomy approach. Recently, with the advancement in endoscopic and endonasal approaches to the cranial base, we have had the opportunity to decompress two patients who had failed craniotomy procedures for optic nerve relief that were operated transnasally and endoscopically with excellent decompression. In experienced hands, the medial and inferior walls of the optic canal can be well decompressed with not only preservation, but also enhancement of deteriorating vision. Overall, the data presented in this report represent a significant approach in the management of optic neuropathy secondary to fibrous dysplasia.

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In this article, the authors are making recommendations about the treatment of optic neuropathy in association with fibrous dysplasia. The natural history data about optic neuropathy and encasement are invaluable, as this is one of the largest series of fibrous dysplasia with follow-up data.

However, the authors provide no details about the techniques of optic nerve decompression. What techniques were used for the decompression, and what methods were used to prevent optic nerve damage during the decompression? It is surprising that the postoperative computed tomographic scans after the optic nerve decompression still showed 100% bony encasement after the decompression. This strongly suggests that the decompression was inadequate and that the results of decompression could be markedly improved in such cases with better surgical technique. I think that asymptomatic patients must be followed with careful radiological and clinical examinations, and optic nerve decompression should be undertaken when there is evidence of either radiographic or clinical progression.

Laligam N. Sekhar
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This is a very interesting report on a relatively rare pathological condition. The authors have collected a large series of patients with a long follow-up period and concluded for the scarce value of a prophylactic optic nerve decompression to prevent visual deterioration. In my experience, the loss of vision that occurred in a few patients was not owing to progressive deterioration, but rather to an abrupt event, namely the expansion of a bone cyst in the proximity the optic pathways (generally the result of a spontaneous intracystic hemorrhage). Also, in our series, subjects with McCune-Albright syndrome tended to experience a more severe clinical course. However, the great majority of our patients showed relatively stable visual deficits for years, with an incidence of progression that diminished significantly after puberty. Consequently, I share the suggestion by the authors to refrain from the prophylactic treatment and consider their advice quite important for our colleagues faced with this rare disease.

Concezio Di Rocco
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In a follow-up to a previous study of 67 optic nerves, the authors have expanded their retrospective series to include 104 patients. Their study included a review of all medical records, endocrine testing, cranial computed tomographic scans, and neuro-ophthalmological examination. In their previous study, they concluded that significant narrowing of the optic canal with fibrous dysplasia was not associated with optic neuropathy (1). This was an important study in that large individual experiences with fibrous dysplasia and the approach to decompression were varied and based more on personal experience than any true literature base. This was true in our own practice, in which we recommend prophylactic decompression based on the assumption that disease progression would result in optic neuropathy. In the current study, the authors demonstrate that, in the majority of cases, vision loss is associated with either growth hormone (GH) excess or the presence of an aneurysmal bone cyst. In 174 optic nerves reviewed in 104 patients, 93% had no evidence of optic neuropathy. They found no case of optic neuropathy in nerves less than 100% encased. Of the 107 optic nerves that were 100% encased, 88% exhibited no evidence of optic neuropathy. Additionally, there were no age differences between groups, indirectly suggesting that it is unlikely that there will be progression resulting in eventual compromise. Sixty-nine percent of the patients with optic neuropathy had either GH excess or an associated aneurysmal bone cyst. Their evidence suggests that, in the absence of the comorbidities of GH excess and/or an associated aneurysmal bone cyst, the baseline rate of optic neuropathy is low. The conclusion is that testing for GH excess and imaging to evaluate for the presence of an aneurysmal bone cyst is an essential component in the care of these patients. The current study establishes that optic nerve decompression should be performed in the presence of progressive optic neuropathy. In the absence of neuropathy, following patients electively may be a safer course of action given the potential compromise associated with surgical decompression.

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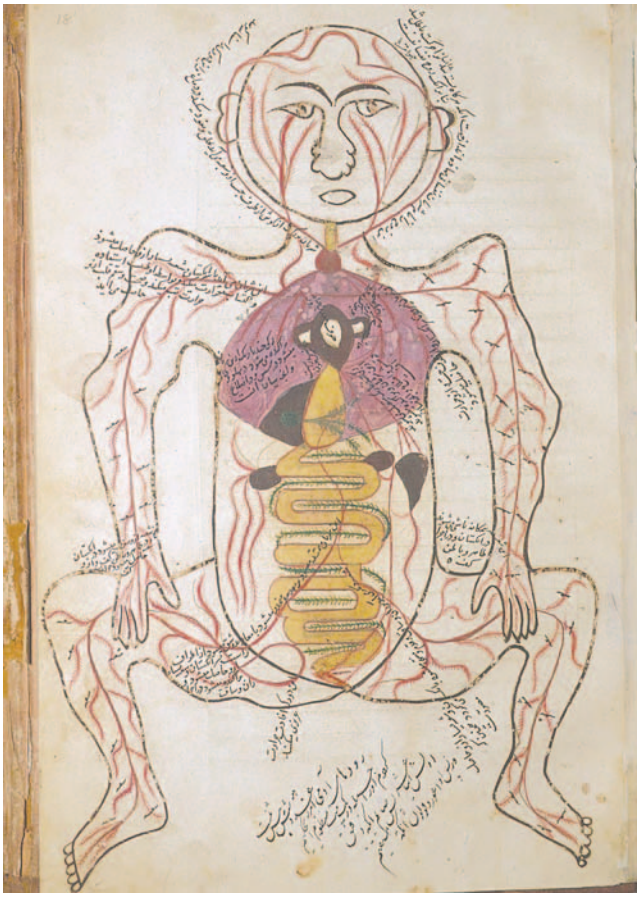
1. Lee JS, FitzGibbon E, Butman JA, Dufresne CR, Kushner H, Wientroub S, Robey PG, Collins MT: Normal vision despite narrowing of the optic canal in fibrous dysplasia. **N Engl J Med** 347:1670–1676, 2002.



Mansur ibn Ilyas, ***Tashrih-i badan-i insan [Anatomy of the Human Body].*** Iran, ca. 1390. (Courtesy of the U.S. National Library of Medicine, National Institutes of Health, Bethesda, Maryland).